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Extracardiac bronchogenic cyst in a 14-year-old female with exercise intolerance

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ABSTRACT

Bronchogenic cysts are foregut-derived cystic malformations of the respiratory tract which may originate anywhere along the developmental pathway of the foregut in an ectopic site. Bronchogenic cysts involving the heart are extremely rare, with evidence of only a few case reports of extracardiac or intracardiac bronchogenic cysts in children. We report a case of an extracardiac bronchogenic cyst in a 14-year-old girl with exercise intolerance, with focus on clinical presentation and diagnostic imaging. Classic findings of bronchogenic cysts on echocardiography are highlighted, in addition to characteristic features that may be identified using cardiac magnetic resonance imaging.

1. Introduction

Bronchogenic cysts are foregut-derived cystic malformations of the respiratory tract [1]. Abnormal or late budding of the embryonic ventral lung bud or the tracheobronchial tree, usually between the 26th and 40th days of gestation, results in an abnormal bud that subsequently differentiates into a fluid-filled, blind-ending pouch [1]. Bronchogenic cysts may originate anywhere along the developmental pathway of the foregut in an ectopic site. Most bronchogenic cysts originate in the mediastinum, while 15% to 20% occur in the lung parenchyma [2]. Bronchogenic cysts involving the heart are extremely rare, with evidence of only a few case reports of extracardiac or intracardiac bronchogenic cysts in children [3–5,7,8,11,12]. We report a case of an extracardiac bronchogenic cyst in a 14-year-old female with exercise intolerance and a heart murmur in order to highlight the unusual clinical presentation and distinguishing characteristics on echocardiogram and cardiac magnetic resonance imaging.

2. Case presentation

A 14-year-old Hispanic girl with history of elevated blood pressure and heart murmur presented to our institution's cardiology clinic for evaluation and clearance for sports. She endorsed inability to participate in competitive running due to severe fatigue 30 min into exercise. She denied other medical history aside from resolved childhood asthma. Her blood pressures had been mildly elevated in the 127–130 mmHg systolic range at the pediatrician's office. Dietary habits were poor with frequent fried food consumption. She denied chest pain, palpitations, dyspnea, syncope, dizziness, or lightheadedness. She denied any fevers, night sweats, or weight loss. Family history was significant for paternal hypertension, otherwise noncontributory for sudden unexplained death, arrhythmias, heart attacks at a young age, connective tissue disorder, or congenital heart disease. On physical exam, the patient's blood pressures were elevated. She had stage 1 hypertension range based on the 2017 American Academy of Pediatrics Clinical Practice Guidelines: right arm 133/75, left arm 131/71, and right leg 173/108 [10]. She was overweight with height of 1.64 m (5′ 4.57″), weight of 74.8 kg (164 lb. 14.5 oz) and body mass index of 27.81 kg/m^2. Pulse was 89 beats per minute, temperature 36.2 °C (97.1 °F), respiratory rate 20 breaths per minute, and oxygen saturation was 100% on room air. She was noted to have a soft grade 1–2/6 systolic, vibratory, low-frequency murmur best heard at the left lower sternal border and apex, louder with supine positioning. There was no rub, click, or gallop noted. The remainder of her physical exam was unremarkable.

Further investigation included electrocardiogram, chest x-ray,

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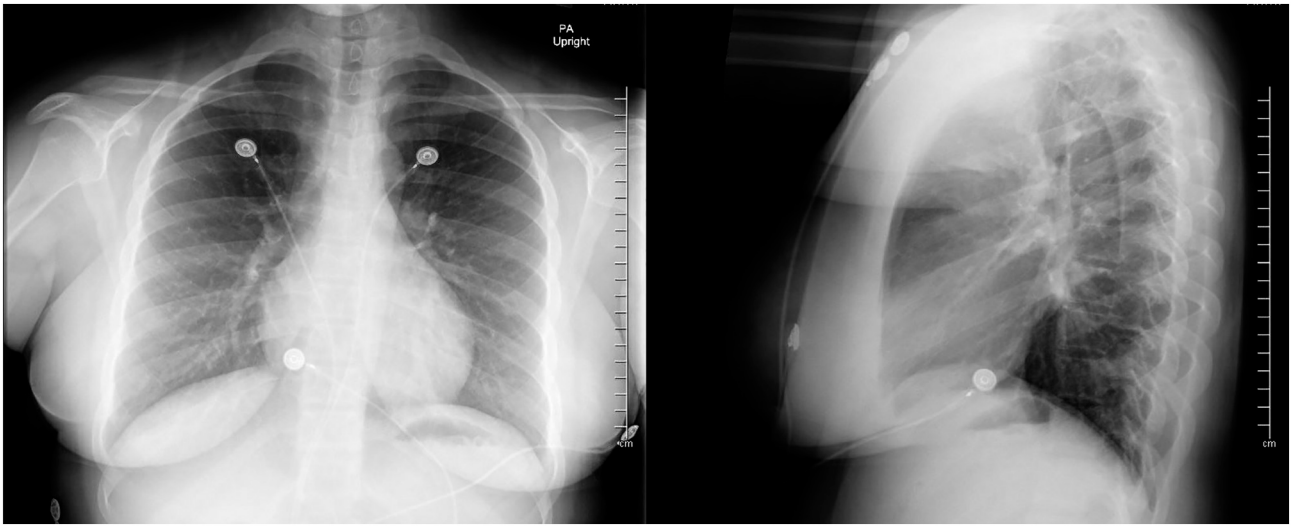

Fig. 1. Normal frontal and lateral views of the chest without any mass or mass effect.

transthoracic echocardiogram, 24 h Holter monitor, and cardiac magnetic resonance imaging. Her electrocardiogram showed normal sinus rhythm without any chamber hypertrophy, ST-T wave changes, ventricular pre-excitation, or QTc prolongation. Chest x-ray showed clear lung fields and normal cardiac silhouette without mass or mass effect (Fig. 1). Echocardiogram showed a 3 × 3.5 cm well-circumscribed, cystic, extracardiac mass compressing the left atrium posteriorly and superiorly without significant obstruction to pulmonary venous return or mitral valve inflow (Fig. 2). Otherwise the rest of the echocardiogram revealed normal intracardiac anatomy, biventricular chamber size and function, no significant left ventricular hypertrophy or arch obstruction, and no intracardiac shunts or valvar dysfunction with normal origin of the proximal coronary arteries (Fig. 2). Differential diagnoses at this time included a benign teratoma, lung tumor, sarcoidosis, thymoma, or mediastinal lymphoma. The patient underwent 24 h Holter monitoring to evaluate for any atrial tachyarrhythmias, which demonstrated normal sinus rhythm throughout, with frequent sinus arrhythmia and good heart rate variability. Cardiac magnetic resonance imaging with and without contrast was performed to better characterize the mass, and showed a large mid/posterior mediastinal, subcarinal, cystic, non-enhancing, extracardiac mass with mass effect on the left atrial wall without invasion of surrounding cardiac structures (Fig. 3). Cardiac magnetic resonance imaging was otherwise normal with normal chamber sizes and biventricular function (Fig. 3). Findings were most consistent with a bronchogenic cyst but did not exclude a foregut duplication cyst, or less likely pericardial cyst or lymphatic malformation. The patient underwent surgical drainage and subtotal resection of the mucous-filled cyst via a right thoracotomy without complications. Due to dense adhesions to the left atrium, a small portion of the cyst was left in situ after fulguration of the lining. Pathology of the cyst wall showed respiratory-type epithelium and intermittent smooth muscle, confirming the diagnosis of bronchogenic cyst.

3. Discussion

In pediatric patients, bronchogenic cysts may cause life-threatening compressive symptoms [1]. However, in adults, bronchogenic cysts are often incidental radiologic findings [1]. This case report presents a 14-year-old overweight girl with complaints of exercise intolerance, hypertension, innocent heart murmur, and structurally normal heart with discovery of an extracardiac mass superior and posterior to the left atrium. Her exercise intolerance, elevated blood pressure, and murmur prompted a cardiac workup, which revealed a cystic structure by echocardiography and cardiac magnetic resonance imaging, later confirmed to be a bronchogenic cyst. The few case reports of children with bronchogenic cysts involving the heart have shown involvement of the right or left atrium [4,5], right or left ventricle [3,6], and ventricular septum in association with a ventricular septal defect [7]. Reported signs and symptoms include an innocent murmur, typically systolic, as in our patient, arrhythmia, cardiac failure, and even cardiac tamponade [3–8]. Other case reports have shown that an underlying bronchogenic cyst may masquerade as chronic or difficult to treat asthma [11,12]. Our patient's history of early childhood asthma may have been related to her undiagnosed bronchogenic cyst. Her exercise intolerance was attributed to a combination of physical deconditioning and mass effect on the left atrium with inefficient filling of the chambers of the heart, especially with exertion. Her murmur was innocent without associated structural heart disease. However, murmurs with bronchogenic cysts have

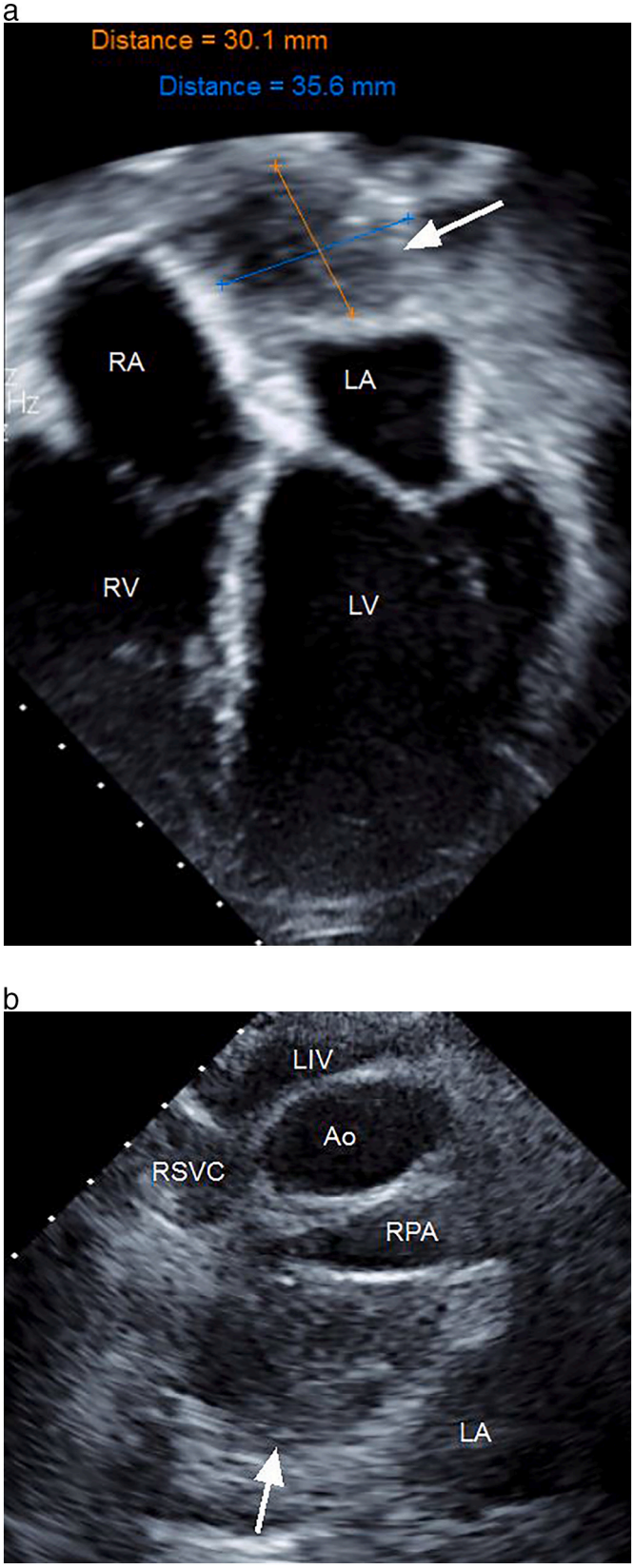

Fig. 2. (a) Apical 4 chamber view demonstrating 3 × 3.5 cm heterogeneous cystic structure (arrow) with mass effect on left atrium. *RA* right atrium, *LA* left atrium, *RV* right ventricle, *LV* left ventricle. (b) Suprasternal short axis view showing bronchogenic cyst (arrow) compressing left atrium. *LIV* left innominate vein, *RSVC* right superior vena cava, *Ao* aorta, *RPA* right pulmonary artery *LA* left atrium.

been reported with congenital anomalies of the heart. Examples include patent ductus arteriosus, atrial septal defect, Tetralogy of Fallot, partial atrioventricular septal defect, and mitral stenosis [13,14]. Furthermore, murmurs may be associated with inlet or outlet obstruction, as in cases of an intracardiac bronchogenic cyst attached to the tricuspid valve or in the right ventricular outflow tract [3,15]. An extracardiac bronchogenic cyst may cause extrinsic compression of the pulmonary artery with associated systolic murmur [16]. There have been no case reports or studies correlating bronchogenic cysts with elevated blood pressure. Our patient's blood pressure on post-surgical follow up visit remained elevated at 130/73. She likely had primary, rather than secondary, hypertension given her positive paternal family history of hypertension and overweight status [10].

This case describes imaging techniques used for diagnosing and treating bronchogenic cysts. Bronchogenic cysts on plain radiographs usually appear as soft-tissue density rounded structures, sometimes with compression of surrounding structures causing air-trapping or a hyperlucent hemithorax. Calcific density material may also be seen [9]. The patient reported here had a normal chest x-ray, which is not uncommon. By cardiac magnetic resonance imaging, a cystic structure is demonstrated with varying signal intensity on T1 images, from low (similar to fluid) to high (due to protein content), or high intensity on T2 imaging due to fluid content [9]. Of note, computed tomography scans are often the initial imaging modality of choice and generally reveal a well-circumscribed spherical or ovoid mass of variable attenuation [1]. However, in the pediatric population, radiation is avoided when possible. Echocardiography is typically only considered when bronchogenic cysts cause cardiac symptoms. Echocardiographic findings may show a hypoechoic cystic structure based on previous case reports or a heterogeneous cystic structure, similar to our patient [3–7]. Diagnostic confirmation of a bronchogenic cyst requires surgical excision and histological examination to show ciliated pseudostratified columnar epithelium of respiratory type, possible areas of squamous metaplasia, and airway components in the cyst wall including cartilage plates, bronchial glands, and smooth muscle. Rarely, nerve and adipose tissues may be seen [1].

4. Conclusion

In summary, bronchogenic cysts, though rare, should be considered in the differential diagnosis of pediatric patients with cardiac symptoms, including exercise intolerance, as extrinsic cardiac compression may be clinically significant, depending on the size and location of the cyst. Echocardiography, followed by cardiac magnetic resonance imaging, is important in the investigation of bronchogenic cysts in the pediatric population if cardiac involvement is suspected. The treatment is surgical excision, and histological examination is used to confirm the diagnosis.

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Declaration of Competing Interest

The authors declare no conflict of interest.

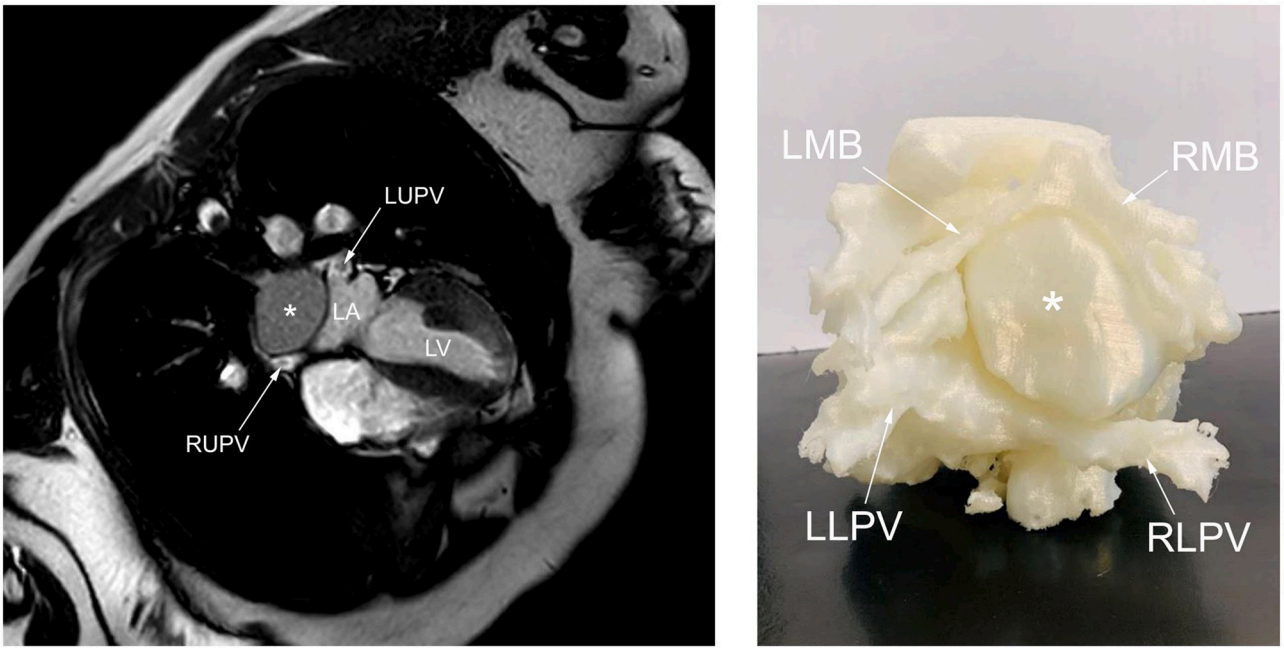

Fig. 3. Magnetic resonance imaging in a standard 4-chamber view and a 3D printed model, demonstrating the posterior cystic mass (*) with compression but no invasion of surrounding cardiac structures. Demonstrated structures are *LUPV* left upper pulmonary vein, *RUPV* right upper pulmonary vein, *LA* left atrium, *LV* left ventricle, *LMB* left mainstem bronchi, *RMB* right mainstem bronchi, *LLPV* left lower pulmonary vein and *RLPV* right lower pulmonary vein.

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